

GUMMATOUS OSTEITIS OF THE SKULL*

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Case Report

A single man aged 65 years was referred to the Department of Venereology in 1949 on account of extensive necrosis of the skull and gangrene of the right foot.

The precipitating factor was an obnoxious smell, attributed to a blocked drain, which was reported by some of his neighbours to the local Sanitary Authority. Through the Medical Officer of Health, the patient was eventually compelled to allow the Sanitary Authority's representative to enter the premises and the source of the smell was soon apparent, that is to say, from the necrotic skull.

History.—According to the patient, in 1905 he suffered from a rash which appeared a few weeks after an acute urethritis. In 1921 he developed a painless ulcer on the right forehead, and similar lesions appeared on the thighs and elbows. These were unsuccessfully treated by the patient himself with ointments and blood mixtures.

In 1926 he reported at the local Venereal Diseases Clinic having developed an enormous ulcer of the scalp extending from the forehead to the occiput and from which, it appears, pieces of necrotic bone were removed. He subsequently attended the local clinic somewhat sporadically until 1935.

Examination.—On his admission to the Department of Venereology, Newcastle, in 1949, examination showed a complete absence of healthy bone above a line encircling the skull, roughly 2 in. above the orbital margins and the ears (Fig. 1). Three-quarters of the vault consisted of brownish-black necrotic bone which crumbled between the fingers, and beneath this and separating it from the brain was a cavity 2 to 2½ in. in depth containing foul-smelling pus. Beneath this there was a layer of flakey, pink or dark brown crusts, the removal of which revealed arterial pulsation in places. The tibiae were grossly thickened but no pain could be elicited on percussion.

There were numerous tissue-paper scars on the upper part of the chest and back and on the arms, and gummatous ulceration of both legs from knees to ankles, accompanied by oedema of the feet. On the distal half of the dorsum of the right foot including the toes, was a black, warty, sodden area with superficial ulceration.

The liver extended two finger-breadths below the costal margin.

Cardiovascular examination revealed systolic and diastolic murmurs in all areas with slight cardiac enlargement, B. P. 142/70.

Examination of the central nervous system showed that Argyll Robertson pupils and slight bilateral exophthalmos were present. Initial deafness was considerably improved by ear syringing, and his sense of taste and smell improved rapidly. The patient had had a speech impediment from infancy. His mental faculties were in some respects above average; arithmetical problems could be calculated with remarkable rapidity and his memory for recent and past events was remarkably good.



FIG. 1.—Syphilitic necrosis of skull.

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Laboratory Investigations.—The blood Wassermann reaction was strongly positive, but the cerebrospinal fluid revealed no abnormalities.

Blood examination initially revealed intense anaemia; but subsequent bi-weekly blood examinations showed a rise in haemoglobin from 6.1 to 11.3 gm./100 ml. followed by a fall to 6.5 gm./100 ml. immediately before death.

Cultures of the pus removed in large quantities from beneath the necrotic skull repeatedly yielded *B. proteus* and *B. pyocyaneus*, both sensitive to streptomycin.

Radiological Reports:

26.8.49. "There is a large area of bone destruction over the vertex of the skull, the frontal bone and both parietal bones being involved. There is an actual defect approximately 4 by 3 ins. with its centre over the bregma. More posteriorly the parietal bones are irregularly eroded, without any new bone formation. The appearance is that of osteitis" (Fig. 2).

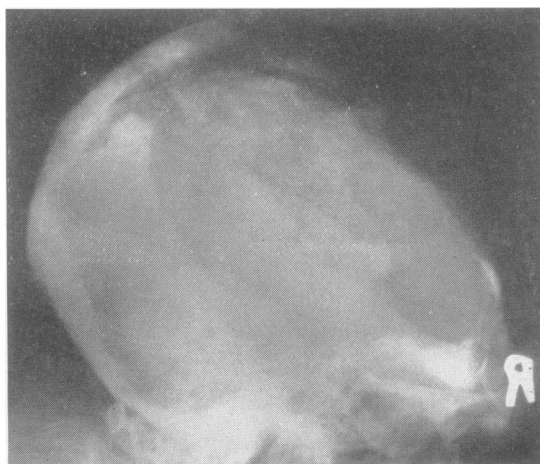


FIG. 2.—X ray of skull.

27.10.49. "There has been some further loss of bone in the posterior part of the vertex, and further involvement of the frontal bone. A sequestrum is forming in the region of the external occipital protuberance. There is more erosion of the right parietal bone extending down through the temporal bone towards the right wing of the sphenoid."

9.5.50. (Fig. 3). "The area of osteitis has increased and now extends anteriorly as far as the supra-orbital ridges and laterally down to the level of the petrous bones. All long bones in both legs show gross periostitis."

The warty growth on the back of the right foot provided a diagnostic problem. While the pathologist could not be dogmatic he did not think that the histology suggested acanthosis nigricans. The clinical picture of chronic lymphoedema surmounted by warty excrescences suggested the condition known as "mossy foot".

Treatment.—Eusol soaks applied to the inside of the skull were later replaced by 1 per cent. Phenoxetol, but neither being effective, dressings of urea and formic iodide were applied with temporary success. Similar dressings

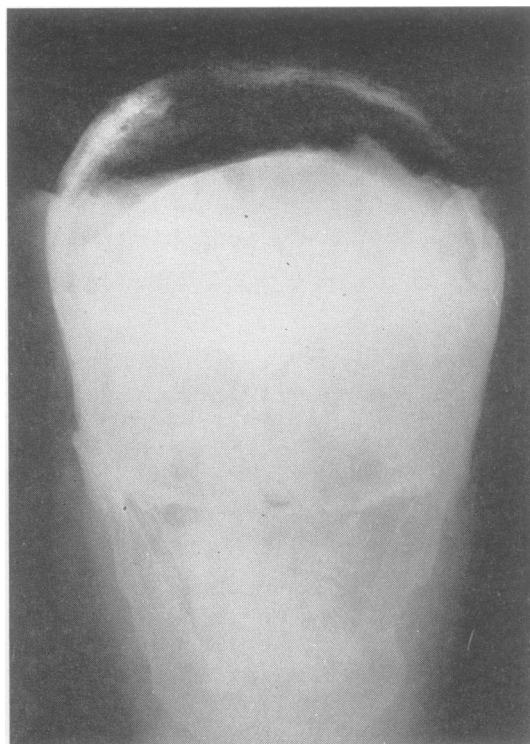


FIG. 3.—X ray of skull 8½ months later.

were applied to the gummatous ulcers on the legs. The lesion on the foot responded to antiseptic dressings and subsequently to the application of vaseline. Medical treatment in the form of Mist. ferri. et ammon. cit., together with a diet rich in proteins, minerals, and vitamins produced a remarkable improvement in the patient's general condition.

Antiluetic Treatment and Subsequent Progress.—Between 1926 and 1935 it would appear, if the available records are correct, that this patient had received three courses of arsenic and 13½ courses of bismuth therapy supplemented periodically by Mist pot. iod. During this time small sequestra would appear to have been removed from the forehead, but apparently the scalp was completely healed by 1935. Subsequent to his reporting at the Newcastle Venereology Department, he received in all two courses of bismuth, 12 gm. streptomycin for the skull infection, and 30 million units of procaine penicillin, and the gummatous lesions responded favourably to this treatment. It is difficult therefore to understand why they had not been averted by the previous bismuth therapy.

The patient's attitude towards his illness, and especially to the skull deformity, was interesting. He was at first reluctant to appear for clinical demonstration, but later this attitude gave way to one of indifference, punctuated by attacks of irritability if the skull dressings were removed for demonstration too frequently. His will to live was



FIG. 4.—Progress of skull necrosis, fully 6 months later.

demonstrated by his determination to retain the tenancy of his Council house, which he did to the day of his death.

His physical and mental condition improved enormously and this might be attributed to dental hygiene as much as anything else. The skull necrosis progressed (compare Figs 1 and 4), but the neurological surgeon advised against surgical interference. His general condition was maintained until approximately one month before death (*i.e.* 9 months after admission to the Venereology Department), when offensive suppuration of the scalp recurred and progressed rapidly in spite of antibiotic therapy, until his relatively sudden death.

Autopsy Report

A *post mortem* examination was made some weeks after death, the body having meanwhile been embalmed. Because of this, autolytic changes were advanced making it difficult to study the histology of the lesions in detail.

Skull.—There was ulceration of the scalp (Figs 5, 6, and 7) from the frontal sinuses to the occiput, with absence of bone over most of the area. The base of the ulcer was composed of crusts up to 1 cm. thick lying on the greatly thickened dura mater, while around the margins of the ulcer there projected a rim of bare necrotic bone. Histologically the crusts were found to consist of hyperkeratotic



FIG. 5.—Hemisection of head. Note thickened dura mater. Most of the covering crust of keratin has been removed from the surface.

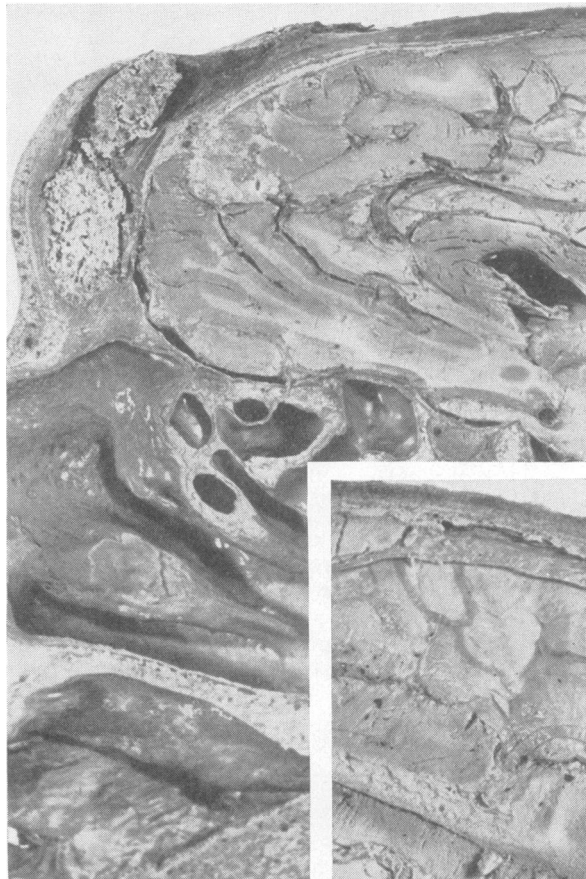


FIG. 6.—Detail of Fig. 5, showing extent of necrosis of frontal bone.

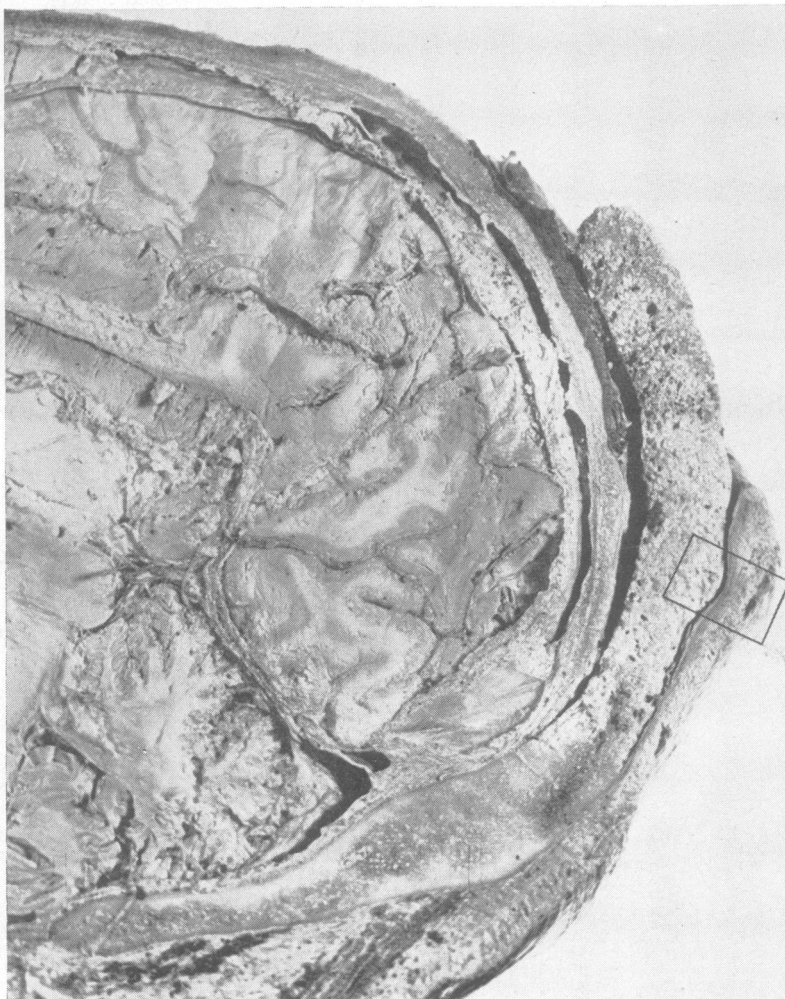


FIG. 7.—Detail of Fig. 5, showing extent of necrosis of occipital bone. The thickening of the dura is also apparent.

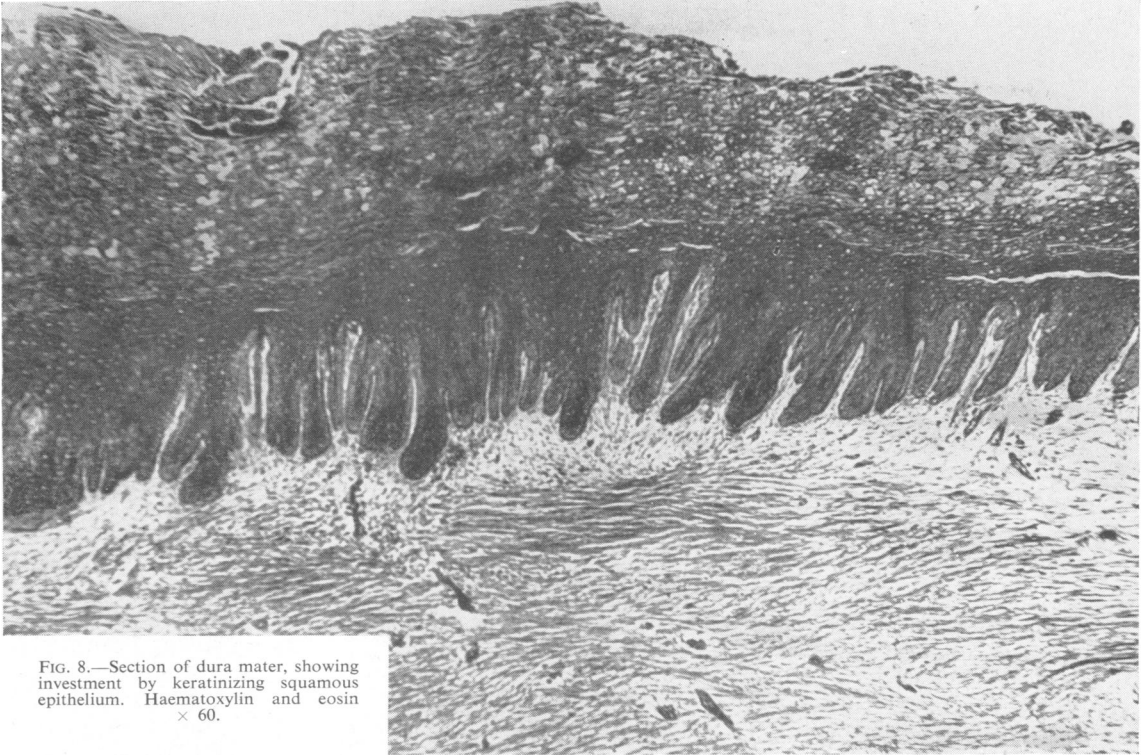


FIG. 8.—Section of dura mater, showing investment by keratinizing squamous epithelium. Haematoxylin and eosin $\times 60$.

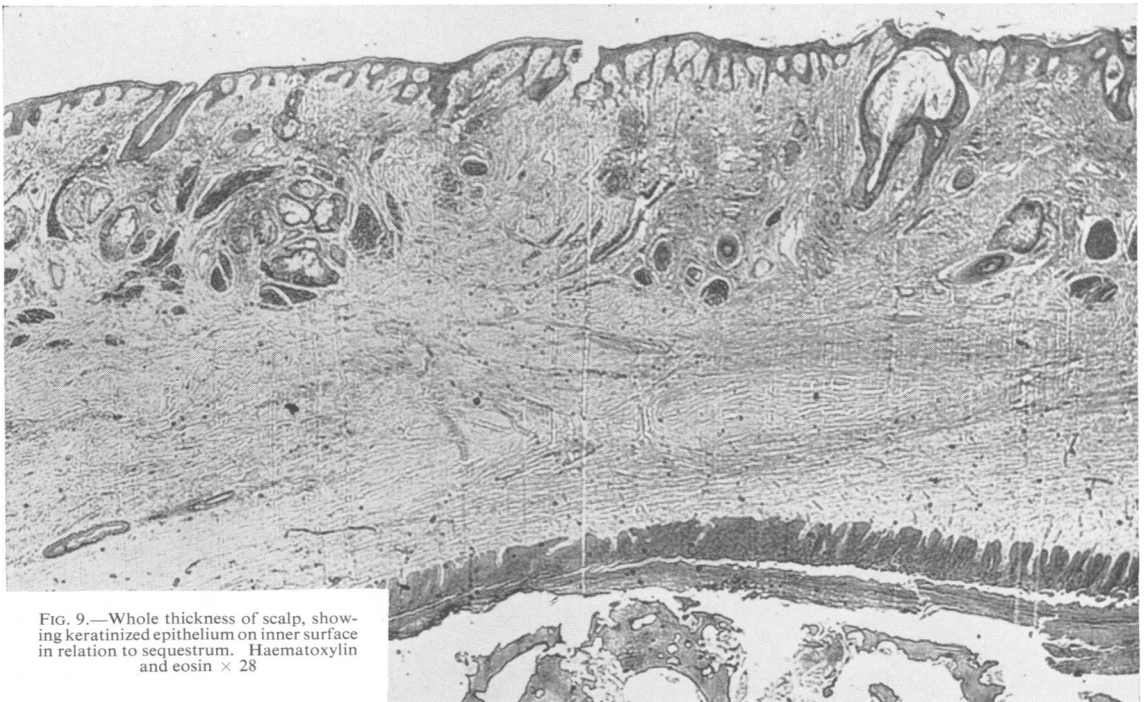


FIG. 9.—Whole thickness of scalp, showing keratinized epithelium on inner surface in relation to sequestrum. Haematoxylin and eosin $\times 28$.

FIG. 10.—Squamous covering of dura, showing apposition of desquamated squames to overlying necrotic bone. Haematoxylin and eosin $\times 65$.

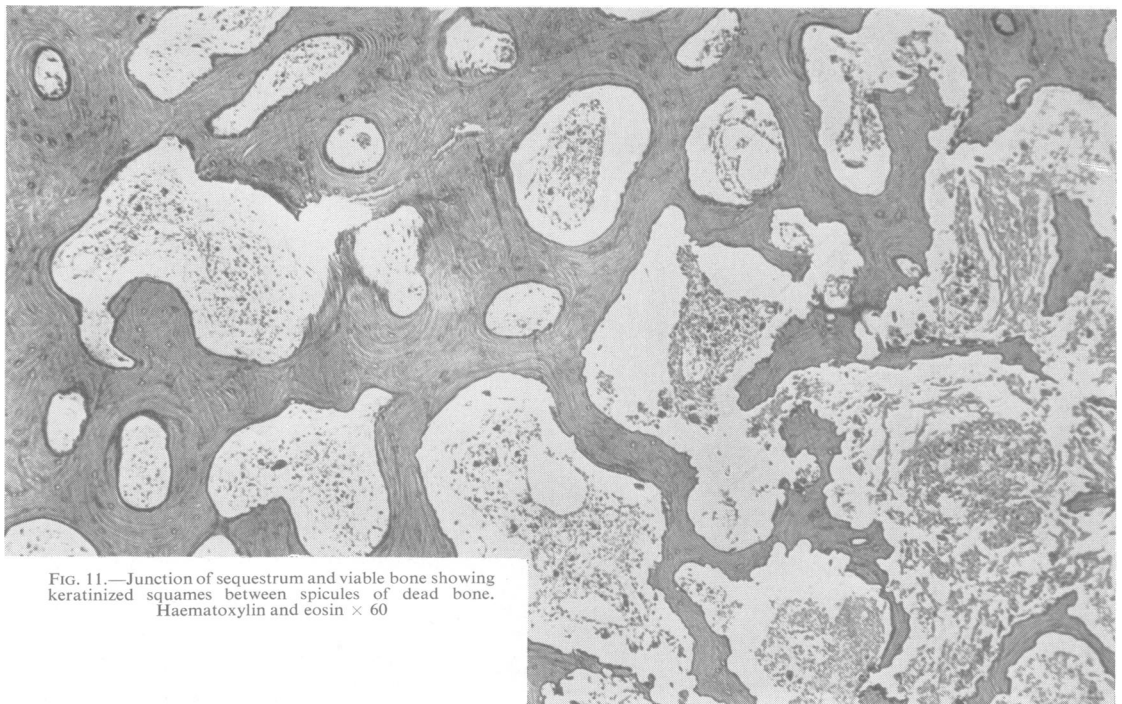
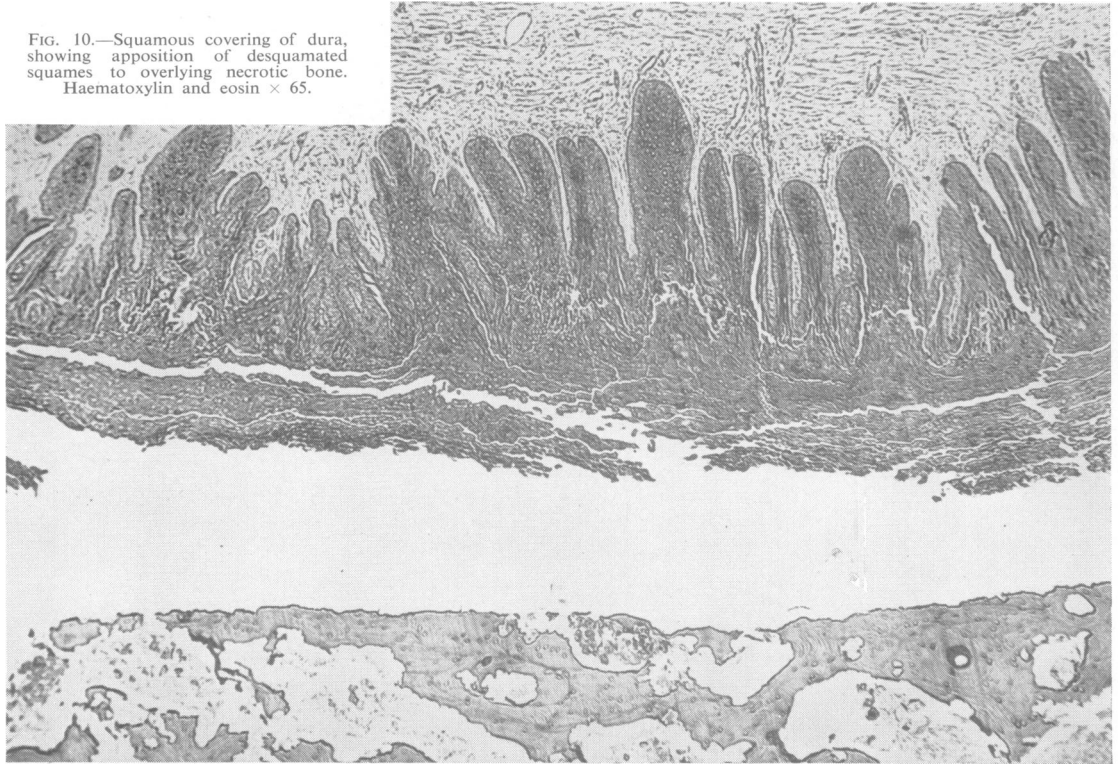


FIG. 11.—Junction of sequestrum and viable bone showing keratinized squames between spicules of dead bone. Haematoxylin and eosin $\times 60$

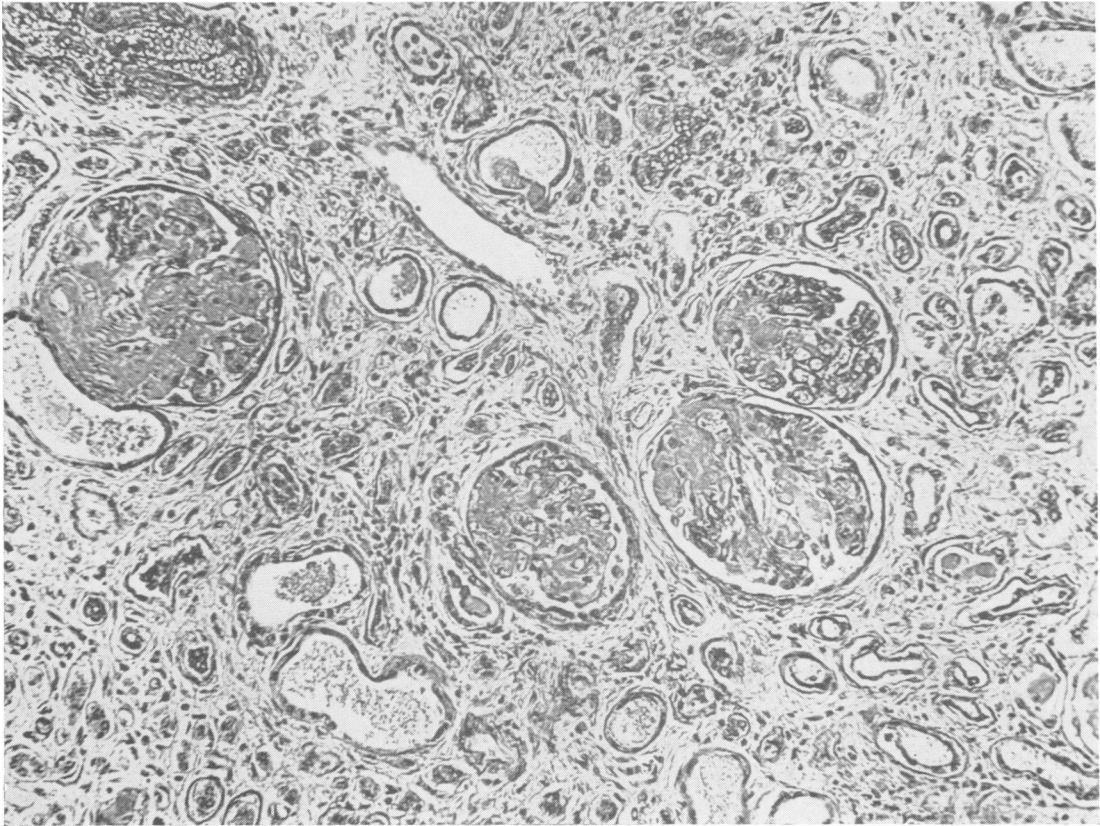


FIG 12.—Section of kidney showing amyloid deposition in glomeruli. Haematoxylin and eosin $\times 180$.

and parakeratotic squamous epithelium (Fig. 8) which formed a thick investment of the exposed part of the dura. The dura itself was thickened by dense fibrous tissue (Fig. 8), but the pia mater and underlying brain tissues were apparently unaffected. At the margin of the ulcer the exposed bone was necrotic but still attached to living bone (Fig. 7). On examining this region in more detail it was found that the skin epithelium was reflected on to the under surface of the scalp so as to separate it from this dead bone (Figs 7 and 9), and also that the dead bone was separated from the underlying dura mater by another layer of squamous epithelium (Fig. 10). There were thus three layers of epithelium with a layer of sequestered bone between the inner two, and an interesting point was that, at the junction of the dead with the living bone, squamous epithelium could be seen penetrating through the interstices to form a connexion (Fig. 11). Thus the whole of the

living tissue was invested by a layer of squamous epithelium.

Sections of the healed gummata on the legs showed periosteal thickening of the long bones. The aorta showed a moderate, apparently inactive, syphilitic mesaortitis.

There was extensive amyloidosis of the kidneys, spleen, liver, adrenals, and bone marrow. In the spleen this had replaced the Malpighian bodies and in the adrenals it had almost entirely replaced the cortex, while in the kidneys it affected the glomerular capillaries (Fig. 12).

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